İntratorasik Desmoplastik Fibroblastoma: Nadir Bir Lokalizasyon

Intrathoracic Desmoplastic Fibroblastoma: A Rare Localization

Desmoplastik Fibroblastoma / Desmoplastic Fibroblastoma

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Özet

Desmoplastik Fibroblastoma (Kollajenöz Fibroma) yakın zamanlarda tanımlanmış, iskelet kası ya da cilt altı dokudan kaynaklanan son derece nadir bir tümördür. Burada, intratorasik yerleşimli bir desmoplastik fibroma olgusu bildirilmiştir. Ellialtı yaşında erkek bir hasta, ağrısız sağ intratorasik kitle ile kliniğimize başvurdu. Fizik muayene, elektrokardiyografi, abdominal ultrasonografi, rutin kan ve biyokimya testleri normaldi. Akciğer radyografisinde sağ alt lobda lobule kontürlü konsolidasyon görüldü. Toraks bilgisayarlı tomografisinde sağ alt hemitoraksı dolduran 10x15 cm boyutunda kitle tespit edildi. Tümör sağ posterolateral torakotomi ile tamamen çıkartıldı ve histopatolojik tanı nadir görülen desmoplastik fibroblastoma olduğunu ortaya koydu. Postoperatif dönem sorunsuz seyretti.

Anahtar Kelimeler

Desmoplastik Fibroblastoma; Kollajenöz Fibroma; Cerrahi

Abstract

Desmoplastic Fibroblastoma (Collagenous Fibroma) is an extremely rare which is a recently described tumor that may arise in the subcutaneous tissue or skeletal muscle. Herewith was reported a case of a Desmoplastic Fibroblastoma with an intrathoracic localization. A 56-year-old man referred with a painless right intrathoracic tumor. Physical examination was normal and electrocardiography, abdominal ultrasonography and routine blood tests were unremarkable. The chest X ray showed consolidation with lobulated contour in right lower zone. Thorax computerized tomography (CT) revealed 10x15 cm mass filling right lower hemithorax. The patient underwent right posterolateral thoracotomy and the tumor was totally removed and histopathologic diagnosis revealed a rare desmoplastic fibroblastoma. Postoperative course was uneventful.

Keywords

Desmoplastic Fibroblastoma; Collagenous Fibroma; Surgery

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Introduction

Desmoplastic Fibroblastoma (DF) is a rare benign soft tissue tumor and recently proposed addition to this group is a tumor first described by Evans et al. [1] as desmoplastic fibroblastoma and later redesignated as collagenous fibroma by Nielsen et al. and Miettinen et al [2,3]. The tumor typically presents as an asymptomatic mass involving the subcutis and skeletal muscle and manifests the most common areas arm, shoulder, lower limb, back, forearm, hands and feet. However, intrathoracic DF is very rare and to our knowledge, no case has been reported in the literature when searched by medline.

Case Report

A 56-year-old man was admitted to our hospital because of a slowly enlarging painless mass in the right lower hemithorax. Physical examination was normal and electrocardiography, abdominal ultrasonography and routine blood tests were unremarkable. The chest X ray showed right lower consolidation with lobulated contour. Thorax computerized tomography (CT) revealed 10x15 cm mass filling right lower hemithorax [figure 1]. Thus, a surgical excision was planned with posterolateral thoracotomy. A right posterolateral thoracotomy through the six intercostal space was performed and revealed a large, encapsulated, elastic and well-described tumor of 10x15-cm diameter. The tumor was attached to the visceral pleura of the right middle lobe and to lateral chest wall. It filled the lower half of the pleural cavity, but was not adherent to any other structure except to fifth rib. The tumor was undiagnosed on frozen section and the preoperative clinical assessment was that it looked malignant.The mass was resected with fifth rib from the pleural cavity, and the right lung expanded completely after the removal of the tumor [figure 2]. The postoperative period was uneventful. The patient was discharged on the 5th postoperative day.

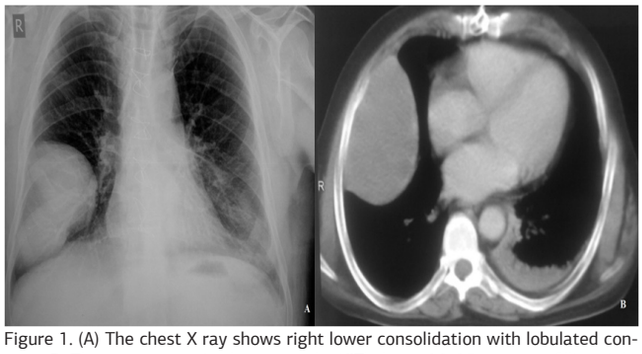

Figure 1. (A) The chest X ray shows right lower consolidation with lobulated contour. (B) Thorax computerized tomography (CT) shows 10x15 cm mass filling right lower thorax

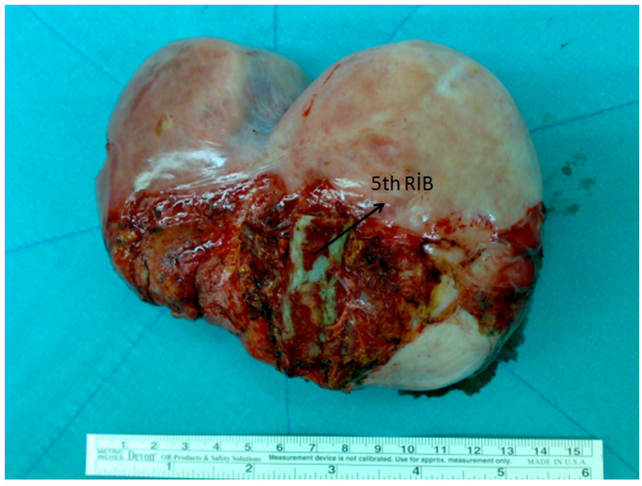

Figure 2. Postoperatively, view of mass with resected 5th rib.

On immunohistochemical examination, there was no immunoreactivity for actin, CD34 and Ki-67. The pathologic examination revealed desmoplastic fibroblastoma.

Discussion

Desmoplastic fibroblastoma, also sometimes known as collagenous fibroma, which is an anatomically ubiquitous but most often subcutaneous lesion occurring in men between the 5th and 7th decades (70%), and rarely in adolescents; only 25% of cases have been diagnosed in women, showing no tendency for local recurrence [4,5]. Generally tumor appears as a well circumscribed, oval or discoid mass localizing the arm, shoulder, lower limb, back, forearm, hands and feet. Its presentation as involvement of the intrathoracic is very rare. Involvement of intrathoracic was not found in the English language medical literature using MEDLINE to the best of our knowledge and a subsequent bibliographic search of all case reports and reviews was performed. The tumor size ranges have been reported from 1 to 20 cm by Miettinen et al [3]. The differential diagnosis includes a variety of benign and low-grade lesions, predominantly fibrous lesions such as neurofibroma, calcifying fibrous pseudotumor, perineurioma, solitary fibrous tumor, elastofibroma, fibroma of tendon sheath, nodular fasciitis, and nuchal fibroma. Cytogenetic analysis in several cases has shown an identical translocation (2;11) (q31;q12) associated with desmoplastic fibroblastoma by Sciot et al and Bernal et al [6,7]. Histologically, these hypocellular tumors consist of stellate and spindle-shaped fibroblast like cells that are widely separated by a collagenous to the fibromyxoid matrix. The plain film findings are nonspecific, and in this case, the lesion consisted of a solid component, suggesting slow, nonaggressive growth. No calcifications were visible on radiography or CT. However we didn't suggest invasive diagnostic tool and we decided exploratory thoracotomy and totally excised the tumor. Since a significant component of the tumor was at least contiguous with the adjacent soft tissue, the intraosseous component in our case probably represents a firmly adhesion of the 5th rib by the tumor.

In summary, collagenous fibroma should be in the differential diagnosis of a well-circumscribed lesion with intraosseous component. Optimal management is conservative excision with functional preservation. Prognosis is excellent and no recurrence is expected.

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